PATIENT REPORT

500 Chipeta Way, Salt Lake City, Utah 84108-1221 phone: 801-583-2787, toll free: 800-522-2787

Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex: Unknown

Specimen Collected: 18-Dec-23 09:36			
Autoimmune Enceph/Dementia Panel, Eserum	Report/Verified: 18-Dec-23 09:54		
Procedure	Result	Units	Reference Interval
Neuronal Antibody (Amphiphysin)	Positive * i1		[Negative]
Purkinje Cell/Neuronal Nuclear IgG Scrn	ANNA Detected * f	1 i2	[None Detected]
NMDA Receptor Ab IgG CBA-IFA, Serum	1:160 * f2 i3		[<1:10]
CASPR2 Ab IgG CBA-IFA Screen, Serum	Detected * t1 i4		[<1:10]
LGI1 Ab IgG CBA-IFA Screen, Serum	m Detected * t2 i5		[<1:10]
CV2 Ab IgG CBA-IFA Screen, Serum	Detected * t3 i6		[<1:100]
AMPA Receptor Ab IgG CBA-IFA Scrn, Serum	Detected * t4 i7		[<1:10]
GABA-BR Ab IgG CBA-IFA Scrn, Ser	Detected * t5 i8		[<1:10]
SOX1 Antibody, IgG by Immunoblot Serum	, High Positive * ⁱ	19	[Negative]
DPPX Ab IgG CBA-IFA Screen, Serum	m Detected * t6 i10		[<1:10]
IgLON5 Ab IgG CBA-IFA Screen, Serum	Detected * t7 i11		[<1:10]
mGluR1 Ab IgG CBA-IFA Screen, Serum	Detected * t8 i12		[<1:10]
Glutamic Acid Decarboxylase Antibody	10.0 H i13	IU/mL	[0.0-5.0]
Neuronal Nuclear Ab (ANNA) IFA Fiter, IgG	Report/Verified: 18-Dec-23 09:54		
Procedure Neuronal Nuclear Ab (ANNA) IFA Titer IgG	Result 1:80 * ⁱ¹⁴	Units	Reference Interval [<1:10]
Neuronal Nuclear Ab IgG, Immunoblot, Ser	Received: 18-Dec-23	3 09:40	Report/Verified: 18-Dec-23 09:54
Procedure Neuronal Nuclear Ab (Hu) IgG,IB Serum	Result , Positive * i15	Units	Reference Interval [Negative]
Neuronal Nuclear Ab (Ri) IgG,IB Serum	, Positive * ⁱ¹⁶		[Negative]
Neuronal Nuclear Ab (Yo) IgG,IB Serum	, Positive * ⁱ¹⁷		[Negative]
Neuronal Nuclear Ab (TR/DNER) IgG,IB	Positive * i18		[Negative]

 $^* = Abnormal, \ \# = Corrected, \ C = Critical, \ f = Result \ Footnote, \ H-High, \ i-Test \ Information, \ L-Low, \ t-Interpretive \ Text, \ @ = Performing \ label{eq:label_equation}$

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ARUP Laboratories

500 Chipeta Way, Salt Lake City, UT 84108

Laboratory Director: Jonathan R. Genzen, MD, PhD

ARUP Accession: 23-352-900114 **Report Request ID**: 18510354

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Patient Age/Sex:

Unknown

AMPA Rptr Ab IgG Titer by CBA-IFA, Ser	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure AMPA Receptor Ab IgG CBA-IFA Titer,Ser	Result 1:160 * ⁱ¹⁹	Units	Reference Interval [<1:10]
CASPR2 Ab IgG Titer by CBA-IFA, Ser	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure CASPR2 Ab IgG CBA-IFA Titer, Serum	Result 1:160 * ¹²⁰	Units	Reference Interval [<1:10]
CV2 Ab IgG Titer by CBA-IFA, Ser	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure CV2 Ab IgG CBA-IFA Titer, Serur	Result n 1:1600 * i21	Units	<pre>Reference Interval [<1:100]</pre>
DPPX Ab IgG Titer by CBA-IFA, Ser	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure DPPX Ab IgG CBA-IFA Titer, Serv	Result .m 1:160 * ¹²²	Units	<pre>Reference Interval [<1:10]</pre>
GABA-B Rptr Ab IgG Titer by CBA-IFA, Ser	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure GABA-BR Ab IgG CBA-IFA Titer,S	Result Ser 1:80 * ⁱ²³	Units	Reference Interval [<1:10]
IgLON5 Ab IgG CBA-IFA Titer, Serum	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure IgLON5 Ab IgG CBA-IFA Titer, Serum	Result 1:160 * ¹²⁴	Units	Reference Interval [<1:10]
LGI1 Ab IgG Titer by CBA-IFA, Ser	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure LGI1 Ab IgG CBA-IFA Titer, Serv	Result 1:80 * ¹²⁵	Units	Reference Interval [<1:10]
mGluR1 Ab IgG CBA-IFA Titer, Serum	Received: 18-Dec-23	09:40	Report/Verified: 18-Dec-23 09:54
Procedure mGluR1 Ab IgG CBA-IFA Titer, Serum	Result 1:160 * ⁱ²⁶	Units	Reference Interval [<1:10]

Interpretive Text

t1: 18-Dec-23 09:36 (CASPR2 Ab IgG CBA-IFA Screen, Serum)

CASPR2 Antibody, IgG is detected. Titer results to follow.

t2: 18-Dec-23 09:36 (LGI1 Ab IgG CBA-IFA Screen, Serum)

LGI1 Antibody, IgG is detected. Titer results to follow.

t3: 18-Dec-23 09:36 (CV2 Ab IgG CBA-IFA Screen, Serum)

CV2 Antibody, IgG is detected. Titer results to follow. Additional charges apply.

t4: 18-Dec-23 09:36 (AMPA Receptor Ab IgG CBA-IFA Scrn, Serum)

AMPAR Antibody, IgG is detected. Titer results to follow.

t5: 18-Dec-23 09:36 (GABA-BR Ab IgG CBA-IFA Scrn, Ser)

GABA-BR Antibody, IgG is detected. Titer results to follow.

t6: 18-Dec-23 09:36 (DPPX Ab IgG CBA-IFA Screen, Serum)

DPPX Antibody, IgG is detected. Titer results to follow.

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phone: 801-583-2787, toll free: 800-522-2787 Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Johathan K. Genzen, Nib, Filib, Onler Medical Onlor

Patient Age/Sex:

<u>Interpretive Text</u>

t7: 18-Dec-23 09:36 (IgLON5 Ab IgG CBA-IFA Screen, Serum)

IgLON5 Antibody, IgG is detected. Titer results to follow.

t8: 18-Dec-23 09:36 (mGluR1 Ab IgG CBA-IFA Screen, Serum)

mGluR1 Antibody, IgG is detected. Titer results to follow.

Result Footnote

f1: Purkinje Cell/Neuronal Nuclear IgG Scrn

Antibodies detected, therefore IFA titer and Immunoblot testing to be performed.

f2: NMDA Receptor Ab IgG CBA-IFA, Serum

Antibodies to NMDA were detected; titer was performed at an additional charge.

The ExTINGUISH Trial (safety and efficacy of Inebilizumab in anti-NMDA receptor encephalitis, NCT04372615) is actively recruiting patients. To learn more, or to refer your patient, call 1-844-427-2465, email ExTINGUISH@hsc.utah.edu, or visit https://neuronext.org/projects/nnlll-extinguish.

<u>Test Information</u>

il: Neuronal Antibody (Amphiphysin)

INTERPRETIVE INFORMATION: Amphiphysin Antibody, IgG

Amphiphysin antibody is present in about 5 percent of patients with stiff-person syndrome and is found variably in other causes of paraneoplastic neurological syndrome (PNS). Amphiphysin antibody is mainly associated with small-cell lung cancer and breast tumors.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i2: Purkinje Cell/Neuronal Nuclear IgG Scrn

INTERPRETIVE INFORMATION: Purkinje Cell/Neuronal Nuclear IgG Scrn

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i3: NMDA Receptor Ab IgG CBA-IFA, Serum

INTERPRETIVE INFORMATION: NMDA Receptor Ab IgG CBA-IFA, Serum

NMDA receptor antibody is found in a subset of patients with autoimmune limbic encephalitis and may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. In addition, positive results have been reported in patients with non-autoimmune phenotypes. A negative test result does not rule out a diagnosis of autoimmune limbic encephalitis. Results should be interpreted in correlation with the patient's clinical history and other

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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex:

Unknown

Test Information

i3: NMDA Receptor Ab IgG CBA-IFA, Serum

laboratory findings. Serum testing should be paired with CSF testing for improved diagnostic sensitivity.

This indirect fluorescent antibody assay utilizes full-length GluN1 transfected cell lines for the detection and semiquantification of NMDA receptor IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i4: CASPR2 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: CASPR2 Ab IgG CBA-IFA Screen,

Serum

Contactin-associated protein-2 (CASPR2) IgG antibody may occur as part of the voltage-gated potassium channel (VGKC) complex antibodies.

The presence of CASPR2 IgG antibody is associated with a wide spectrum of clinical manifestations, including acquired neuromyotonia, limbic encephalitis, painful neuropathy, and Morvan syndrome. Tumors such as thymoma, small cell lung cancer, and other rarer tumors may occur. The full-spectrum of clinical disorders and tumors associated with the CASPR2 IgG antibody continues to be defined. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes CASPR2 transfected cell lines for the detection and semiquantification of the CASPR2 IqG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i5: LGI1 Ab IgG CBA-IFA Screen, Serum INTERPRETIVE INFORMATION: LGI1 Ab IgG CBA-IFA Screen, Serum

Leucine-rich, glioma-inactivated 1 protein (LGI1) IgG antibody may occur as part of the voltage-gated potassium channel (VGKC) complex antibodies.

The presence of LGI1 IgG antibody is mainly associated with limbic encephalitis, hyponatremia, and myoclonic movements. LGI1 IgG antibody is rarely associated with tumors but may occur infrequently in Morvan syndrome, neuromyotonia, and idiopathic epilepsy. The full-spectrum of clinical disorders associated with the LGI1 IgG antibody continues to be defined. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

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Patient Age/Sex:

Unknown

Test Information

i5: LGI1 Ab IgG CBA-IFA Screen, Serum

This indirect fluorescent antibody assay utilizes LGI1 transfected cell lines for the detection and semiquantification of the LGI1 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i6: CV2 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: CV2 Ab IgG CBA-IFA Screen, Serum

CV2 antibodies aid in discriminating between chronic paraneoplastic neurological disorder (PND) and other inflammatory disorders of the nervous system. Anti-CV2 is associated with small-cell lung cancer and thymoma. A negative test result does not rule out a diagnosis of autoimmune neurologic disease. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes CV2 transfected cell lines for the detection and semiquantification of the CV2 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i7: AMPA Receptor Ab IgG CBA-IFA Scrn, Serum

INTERPRETIVE INFORMATION: AMPA Receptor Ab IgG CBA-IFA Scrn,

Serum

Alpha-amino-3-hydroxy-5-methyl-4-isoxazoleproprionic acid receptor (AMPAR) antibody is found in a subset of patients with autoimmune limbic encephalitis and may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. A negative test result does not rule out a diagnosis of autoimmune encephalitis. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes AMPAR transfected cell lines for the detection and semiquantification of AMPAR IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i8: GABA-BR Ab IgG CBA-IFA Scrn, Ser

INTERPRETIVE INFORMATION: GABA-BR Ab IgG CBA-IFA Scrn, Ser

*_Abnormal #_Corrected C_Critical f_Pocult Footnote H High i Toot Information L Low + Interpretive Text @_Porforming lab

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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex:

Unknown

Test Information

i8: GABA-BR Ab IgG CBA-IFA Scrn, Ser

Gamma-amino butyric acid receptor, type B (GABA-BR) antibody is found in a subset of patients with autoimmune epilepsy and other autoimmune neurologic phenotypes; it may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. A negative test result does not rule out a diagnosis of autoimmune neurologic disease. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes GABA-BR transfected cell lines for the detection and semiquantification of GABA-BR IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i9: SOX1 Antibody, IgG by Immunoblot, Serum

INTERPRETIVE INFORMATION: SOX1 Antibody, IgG by Immunoblot,

Serum

SOX1 antibody is detected in patients with Lambert-Eaton myasthenic syndrome (LEMS) and in patients with paraneoplastic cerebellar degeneration (PCD), paraneoplastic and nonparaneoplastic neuropathy. SOX1 antibody is associated with small cell lung cancer. A negative test result does not rule out a diagnosis of LEMS or other causes of paraneoplastic neurological syndrome.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

il0: DPPX Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: DPPX Ab IgG CBA-IFA Screen, Serum

DPPX antibody is found in a subset of patients with autoimmune encephalitis, and is often associated with prodromal gastrointestinal symptoms and unintentional weight loss. It may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. A negative test result does not rule out a diagnosis of autoimmune neurologic disease. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes DPPX transfected cells for the detection and semiquantification of the DPPX IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex: Unknown

Test Information

ill: IgLON5 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: IgLON5 Ab IgG CBA-IFA Screen,

Serum

IgLON Family Member 5 (IgLON5) antibody is found in a subset of patients with autoimmune encephalitis or other autoimmune neurologic/neurodegenerative disorders and may occur with or without associated tumor. A negative test result does not rule out a diagnosis of an autoimmune neurologic disorder. Interpretation of any antineural antibody test requires clinical correlation.

This indirect fluorescent antibody assay utilizes IgLON5 transfected cell lines for detection and semi-quantification of IgLON5 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

i12: mGluR1 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: mGluR1 Ab IgG CBA-IFA Screen,

Serum

Metabotropic glutamate receptor 1 (mGluR1) antibody is found in a subset of patients with autoimmune cerebellar ataxia or autoimmune encephalitis and may occur with or without associated tumor. A negative test result does not rule out a diagnosis of autoimmune cerebellar ataxia or limbic encephalitis. Interpretation of any antineural antibody test requires clinical correlation.

This indirect fluorescent antibody assay utilizes mGluR1 transfected cell lines for detection and semi-quantification of mGluR1 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

il3: Glutamic Acid Decarboxylase Antibody

INTERPRETIVE INFORMATION: Glutamic Acid Decarboxylase Antibody

A value greater than 5.0 IU/mL is considered positive for Glutamic Acid Decarboxylase Antibody (GAD Ab). This assay is intended for the semi-quantitative determination of the GAD Ab in human serum. Results should be interpreted within the context of clinical symptoms.

i14: Neuronal Nuclear Ab (ANNA) IFA Titer IgG

INTERPRETIVE INFORMATION: Neuronal Nuclear Ab (ANNA) IFA Titer IgG

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug

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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex:

Unknown

Test Information

i14: Neuronal Nuclear Ab (ANNA) IFA Titer IgG

Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i15: Neuronal Nuclear Ab (Hu) IgG, IB, Serum

INTERPRETIVE INFORMATION: Neuronal Nuclear Ab IgG,

Immunoblot, Ser

This test detects IgG antineuronal antibodies to Hu, Ri, Yo and Tr (DNER) antigens.

Antineuronal antibodies serve as markers that aid in discriminating between a true paraneoplastic neurological disorder (PND) and other inflammatory disorders of the nervous system. Anti-Hu (antineuronal nuclear antibody, type I) is associated with small-cell lung cancer. Anti-Ri (antineuronal nuclear antibody, type II) is associated with neuroblastoma in children and with fallopian tube and breast cancer in adults. Anti-Yo (anti-Purkinje cell cytoplasmic antibody) is associated with ovarian and breast cancer. Anti-Tr(DNER) is associated with Hodgkin's lymphoma.

The presence of one or more of these antineuronal antibodies supports a clinical diagnosis of PND and should lead to a focused search for the underlying neoplasm.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i16: Neuronal Nuclear Ab (Ri) IgG, IB, Serum

INTERPRETIVE INFORMATION: Neuronal Nuclear Ab (Ri) IgG, IB,

Serum

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i17: Neuronal Nuclear Ab (Yo) IgG, IB, Serum

INTERPRETIVE INFORMATION: Neuronal Nuclear Ab (Yo) IgG, IB,

Serum

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i18: Neuronal Nuclear Ab (TR/DNER) IgG, IB

INTERPRETIVE INFORMATION: Neuronal Nuclear Ab (TR/DNER)

IgG, IB

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Unknown

Test Information

i19: AMPA Receptor Ab IgG CBA-IFA Titer, Ser

INTERPRETIVE INFORMATION: AMPA Receptor Ab IgG CBA-IFA

Titer, Ser

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i20: CASPR2 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: CASPR2 Ab IgG CBA-IFA Titer, Serum

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i21: CV2 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: CV2 Ab IgG CBA-IFA Titer, Serum

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i22: DPPX Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: DPPX Ab IgG CBA-IFA Titer, Serum

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i23: GABA-BR Ab IgG CBA-IFA Titer, Ser

INTERPRETIVE INFORMATION: GABA-BR Ab IgG CBA-IFA Titer, Ser

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i24: IgLON5 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: IgLON5 Ab IgG CBA-IFA Titer, Serum

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

i25: LGI1 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: LGI1 Ab IgG CBA-IFA Titer, Serum

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Patient Age/Sex:

Unknown

Test Information

i25: LGI1 Ab IgG CBA-IFA Titer, Serum

> This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i26: mGluR1 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: mGluR1 Ab IgG CBA-IFA Titer, Serum

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